Journal of Cardiology Cases (2012) **5**, e20–e22

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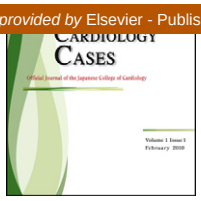

Case Report

Anomalous origin of right coronary artery from pulmonary artery presenting as chest pain in a young man

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Received 8 July 2011; received in revised form 29 August 2011; accepted 6 September 2011

KEYWORDS
Anomalous coronary artery;
Chest pain;
ARCAPA

Summary Isolated anomalous origin of right coronary artery is a rare developmental anomaly which is mostly asymptomatic and is discovered incidentally. We present a case of a 21-year-old male who presented with chest pain and was found to have anomalous origin of right coronary artery from pulmonary artery for which he underwent prompt surgical correction.

Case report

A 21-year-old African American male presented with dull central chest pain which started 12 h previously, was gradually progressing, and was associated with generalized weakness. Review of systems was pertinent for reduced exertional capacity due to shortness of breath. Family history was negative for sudden cardiac death and premature coronary heart disease. He worked as bus driver, had a predominantly sedentary lifestyle, and smoked marijuana occasionally.

Assessment

On examination he appeared in discomfort with blood pressure of 157/102 mmHg and 154/100 mmHg in both arms and bilaterally symmetrical pulse of 107/minute. There was no jugular vein distention or carotid bruit in neck. Cardiac examination was unremarkable except for tachycardia and breath sounds were equal bilaterally. Extremities were free of edema.

Electrocardiography showed sinus tachycardia with T wave inversion and borderline ST segment elevation in lateral leads (Fig. 1). Chest X-ray showed normal heart and mediastinum size (Fig. 2). Laboratory results showed elevated cardiac enzymes i.e. creatine kinase-MB 37.1, cardiac troponin I 8.24, with normal renal and liver function, white blood cell count was 8.4, and erythrocyte sedimentation rate (ESR) was 74. Because of concern of aortic dissection an urgent bedside trans-thoracic echocardiogram was done (as it happened to be instantly available). It showed inferior wall hypokinesia with no evidence of aortic root dilatation or dissection flaps. Urine toxicology was negative for cocaine.

Urgent diagnostic cardiac catheterization of left coronary circulation showed dilated and tortuous left coronary artery without any obstructive pathology. After a short delay there was opacification of the right coronary artery via collateral which was seen to be draining into the pulmonary artery. This established the diagnosis of anomalous origin of right

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1878-5409/$ – see front matter
doi:10.1016/j.jccase.2011.09.007

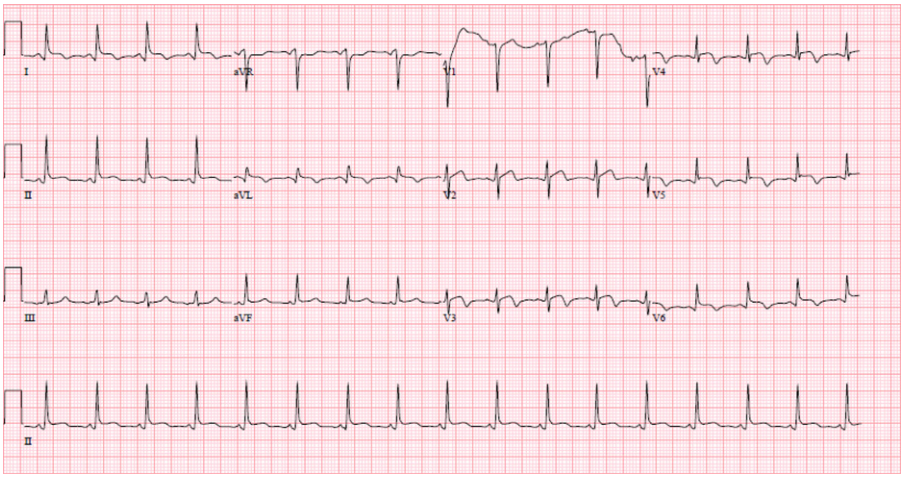

Figure 1 Electrocardiogram showing borderline ST elevation and T wave inversion predominantly in lateral leads.

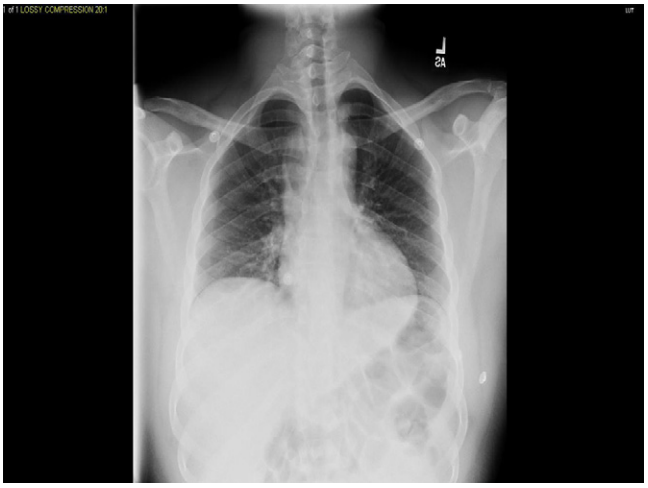

Figure 2 Chest X-ray showing normal heart and mediastinal size.

coronary artery from pulmonary artery (ARCAPA) (Fig. 3 and Video 1).

Diagnosis and management

With such a presentation in an emergent situation several differentials were on top of our list: (1) Acute coronary syndrome — Even though he fulfilled all the criteria for acute coronary syndrome, it was less likely because of his young age (2) Aortic dissection (AD) was strongly considered despite the absence of typical signs and symptoms of tearing chest pain, pulse deficit, and aortic insufficiency. Transthoracic echocardiogram showed no evidence of aortic root dilatation, regurgitation, or dissection flap. It did not completely rule out AD, but it ruled out high-risk features and complications such as pericardial effusion and tamponade. Meredith et al. [1] reported under-utilization of transthoracic echocardiogram in emergency situations in their study. (3) Perimyocarditis was also a possibility and the patient did have elevated ESR, however, echocardiogram showed segmental inferior wall motion abnormality without any effusion. Intraoperatively there were no signs of any pericardial exudates. Moreover, the patient's troponins were too high to be simply explained by perimyocarditis. We did not perform cardiac magnetic resonance imaging as the patient received operative intervention on the following day. (4) Transient coronary flow reduction or coronary spasm was also a diagnostic possibility but the patient did not give any history of it and urine toxicology screen was negative for any illicit drugs. Coronary angiography did not show any evidence of obstruction. (5) Undiagnosed coronary anomaly was considered a strong possibility. The patient's unremarkable medical and family history made any adult congenital heart disease less likely. In retrospect, the patient's limited functional capacity and sedentary lifestyle might have been due to the underlying disease. Diagnosis of ARCAPA was confirmed by coronary angiography. The patient underwent surgical reimplantation of the RCA to aortic root establishing dual ostial circulation.

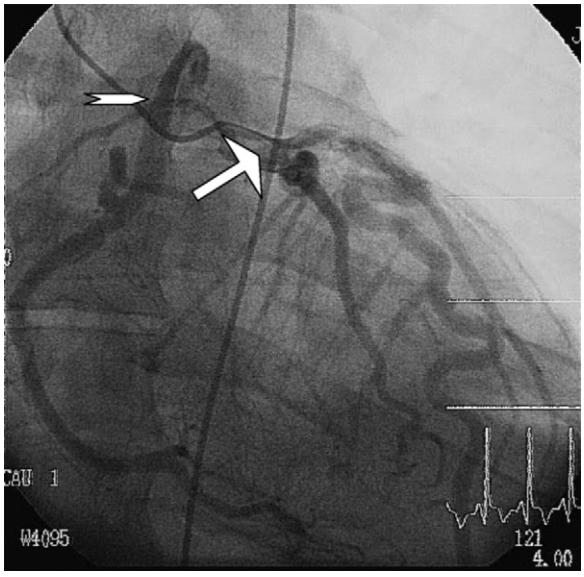

Figure 3 Coronary angiogram of left coronary circulation via left coronary ostia (arrow) showing drainage of blood into pulmonary artery via right coronary artery (arrowhead) leading to phenomenon of intercoronary steal.

Discussion

Estimates of population prevalence of coronary anomaly are based on angiographic studies of adult population and range from 0.6—1.3% [2—11]. It does not estimate true prevalence for two reasons. Firstly, many of the anomalies are fatal in infancy and secondly, compared to multi-detector computed tomography, conventional coronary angiogram may underestimate the prevalence [10,12].

There have been various classifications proposed based on anatomy, angiographic, and hemodynamic findings. According to Greenberg et al. [6] major anomalies leading to abnormal myocardial perfusion are anomalous origin from pulmonary artery, origin of coronary artery from opposite or non-coronary sinus and an abnormal course, myocardial bridging, and coronary artery fistula.

Anomalous left coronary artery from pulmonary artery (ALCAPA) is more common than ARCAPA [3,8,13,14] and is mostly fatal in infancy. Incidence of ARCAPA is estimated to be 0.002% [11]. Isolated ARCAPA is mostly asymptomatic in early life [2,13,15], 25—30% cases are associated with congenital disease most commonly aortopulmonary window and tetralogy of Fallot [16].

The time of onset and severity of symptoms depends on the type of anomaly, direction of blood flow in the anomalous vessel, and extent of collateralization [13]. Normally, there is a retrograde flow in the anomalous artery due to pressure difference between systemic and pulmonary circulation leading to intercoronary steal phenomena. Any increase in oxygen demand leads to exhaustion of the physiologic reserve resulting in ischemia or infarction [17].

Common presenting signs and symptoms of ARCAPA are murmur, angina, dyspnea on exertion, congestive heart failure, and sudden cardiac arrest. In a study by Eckart et al. of 126 nontraumatic sudden deaths in young adults, cardiac abnormality was found in 64 cases (51%), with coronary artery abnormalities being the most common cardiac abnormality (39 of 64 patients) [18]. Because of the risk of sudden death and improvement in operative morbidity, surgical correction with reestablishment of dual coronary ostial circulation is recommended even in asymptomatic patients [13,19].

Appendix A. Supplementary data

Supplementary data associated with this article can be found, in the online version, at doi:10.1016/j.jccase.2011.09.007.

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